diagnosis of Wilson's disease is not straightforward as there are more than 190, usually rare, mutations.[2] None the less, haplotype analysis can provide a definitive method for establishing carrier status in first degree relatives as in subject III:2 (fig 2).

Finally, one recent study suggested that dystonic tremor is often misdiagnosed as essential tremor, even by neurologists[5]; thus it is vital that the possibility of Wilson's disease is considered in every patient under the age of 50 with a progressive movement disorder, even where the family history does not suggest a recessive mode of inheritance.

1 Walshe JM. Wilson's disease. In: Vinken PJ, Bruyn GW, Klawans HL, eds. *Handbook of neurology*. New York: American Elsevier, 1986:223–38.
2 Kenney S, Cox D. *Wilson disease mutation database*. (http://www.medgen.med.ualberta.ca/database.html, accessed 28 August 2000).
3 Quinn NP, Marsden CD. Coincidence of Wilson's disease with other movement disorders in the same family. *J Neurol Neurosurg Psychiatry* 1986;**49**:221–2.
4 Steindl P, Ferenci P, Dienes HP, *et al*. Wilson's disease in patients presenting with liver disease: a diagnostic challenge. *Gastroenterology* 1997;**113**:212–18.
5 Schrag A, Muenchau A, Bhatia KP, *et al*. Overdiagnosis of essential tremor. *Lancet* 1999;**353**:1498–9.

NEUROLOGICAL PICTURE

"The other" Babinski's sign: paradoxical raising of the eyebrow in hemifacial spasm

Joseph Babinski is famous for his description, in 1896, of the abnormal plantar reflex as an indicator of dysfunction in the pyramidal tract. After the works of Brissaud and Meige, his contribution to description of hemifacial spasm is much less well known.

He reported for the first time paradoxical syncinesis in hemifacial spasm in a lecture given at the Société Neurologique de Paris on 6 April 1905.[1] "The most singular is the following: when orbicularis oculi contracts and the eye closes, the internal part of the frontalis contracts at the same time . . . the eyebrow rises during eye occlusion . . . this set of occurrences is impossible to reproduce by will . . ."

From these observations, Babinski concluded that hemifacial spasm is neither the result of a psychological problem nor of a cortical lesion, but instead is due to a lesion that affects directly the facial nerve.

This "other" Babinski's sign can, occasionally, be useful in distinguishing hemifacial spasm from other craniofacial movement disorders.

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1 Babinski J. Hémispasme facial périphérique. *Nouvelle iconographie de la Salpétrière* 1905;**18**:418–23.

Figure 1 Left hemifacial spasm. Paradoxical raising of the eyebrow as the eye closes (the "other" Babinski's sign) (with permission).